Cardiac Catheterization in Adult Congenital Heart Disease

A Preliminary Report

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SUMMARY

The diagnosis of uncomplicated ventricular septal defect and uncomplicated patent ductus arteriosus presents few hazards. Differential diagnosis of atrial septal defect and of transposed pulmonary veins is technically more difficult. The complex nature of most types of cyanotic congenital heart disease requires the combined use of catheterization, determination of circulation times, ventilation studies, and possibly angiocardiography.

Reports of eight cases in which cardiac catheterization was carried out illustrate the common forms of acyanotic and cyanotic heart disease and the factors in diagnosis.

THE purpose of this presentation is to illustrate the value of cardiac catheterization in study of congenital heart disease in adults.

MATERIAL AND METHOD

For purposes of report, eight cases of adult congenital heart disease in which catheterization was carried out at the Los Angeles County General Hospital during the years 1948 and 1949 were selected. Routine clinical work-up was followed by electrocardiography and fluoroscopy in each case. Catheterization of the heart was then carried out by the method of Cournand and co-workers.[2] Oxygen content of blood samples was measured by the Van Slyke method. The intracardiac pressures were measured by a variety of techniques including simple saline manometer, electromanometer, and by multichannel recording with strain gauge manometers.

The diagnostic value of cardiac catheterization depends primarily on the correlation of three separate kinds of information—pressure, oxygen content, and catheter position. Normal "standards" and criteria for abnormal hearts have not been universally accepted. The standards probably will change with further refinements in technique. Technical difficulties during the procedure may materially alter results. Illustrative are tachycardia induced by the "touch" effect of the tip of the catheter against the right ventricular wall, a febrile reaction during the procedure, or a change in heart rate such as may be caused by excitement. Physiologic determinations (sampling, pressures, etc.), should be made as rapidly as possible to help circumvent these problems. The technical difficulties in Van Slyke determinations and in pressure determinations appear less variable.

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I. NORMAL VALUES

1. *Atrium and Great Veins*

Normally, the superior and inferior vena cava contain blood of low oxygen content; that of the inferior vena cava is usually higher than that of the superior vena cava. Under resting conditions in the normal patient, oxygen content of superior and inferior cava blood lies between 12 and 14 volumes per cent. This presupposes the normal A-V difference of 4 to 5 volumes per cent and a normal hemoglobin content of 15 grams per 100 cc. of blood. Inferior cava blood may have oxygen content as much as 2 volumes per cent higher than does superior cava blood. This difference is presumably due to the high oxygen content of renal venous blood.

The pressure in the great veins is normally the lowest in the cardiovascular system, and varies somewhat with normal respiratory excursion. It consists of a, c, and v waves, the familiar venous pulse.

Oxygen content of right atrial blood is representative of a mixture of superior and inferior cava blood, together with the extremely unsaturated coronary sinus blood. The oxygen content of the latter is usually around 4 volumes per cent with the patient in the resting state. Right auricular oxygen content, then, may vary considerably, depending upon the proximity of the tip of the catheter to the opening of the coronary sinus, to the superior vena caval stream or to the inferior vena caval stream.

2. *Right Ventricle*

The form of the pressure wave within the right ventricle is exactly similar in contour in the normal resting state to that of the left ventricle, but the peak systolic pressure is approximately one-fourth that of the left side of the circulation, usually from 20 to 25 mm. of mercury. The form of the curve is that of an inverted "U" with rather similar ascending and descending limbs. The pressure during diastole falls to a level of 0 to 2 mm. of mercury. Oxygen content is that of a mixture of superior and inferior cava blood and blood from the coronary sinus. It may be slightly higher or slightly lower than the content in the right auricle by a difference of 1 volume per cent, depending upon the site from which the sample from the right atrium is taken.

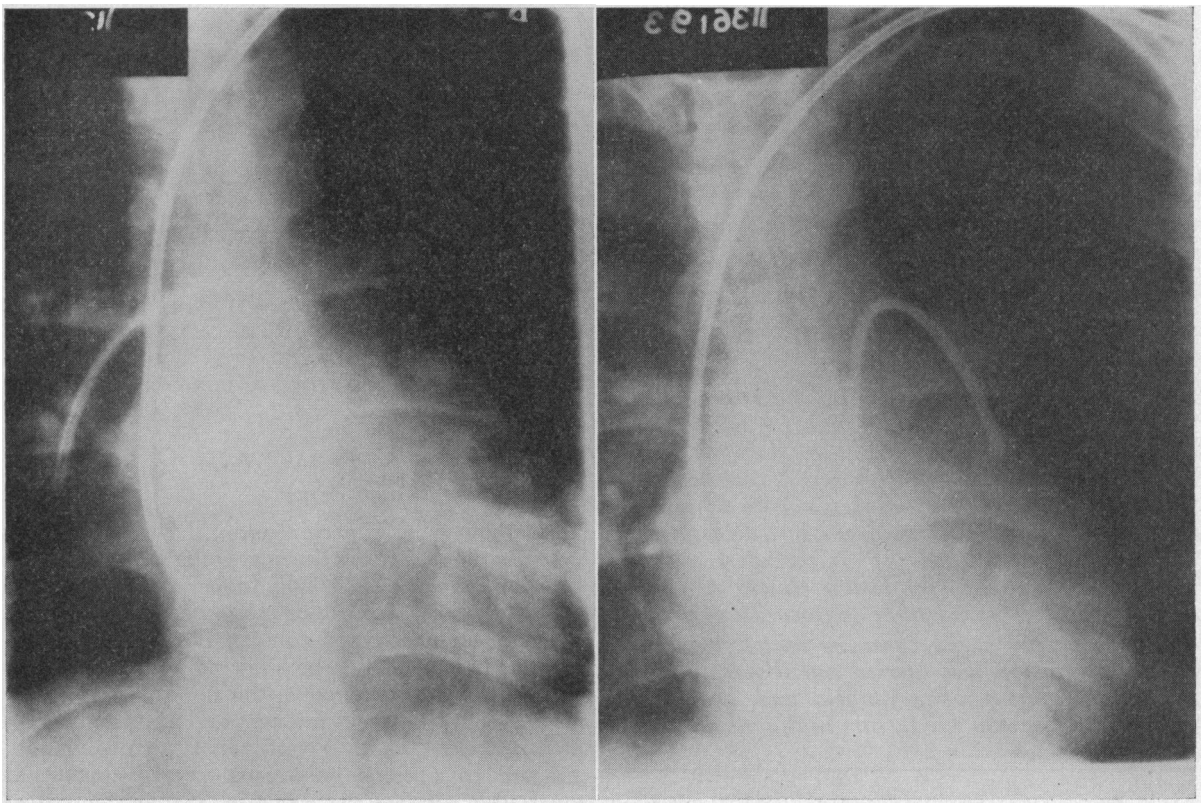

Figure 1.—*Left,* catheter in right pulmonary artery. *Right,* catheter in left pulmonary artery.

3. *Pulmonary Artery*

Normal pulmonary artery pressure curves closely resemble those of the aorta in contour and are approximately one-fourth as high in both systolic and diastolic values. Peak systolic pulmonary artery pressure always is identical to peak right ventricular systolic pressure. The oxygen content in the pulmonary artery should not vary from that of the right ventricle by more than 0.5 volumes per cent. Catheter positions in right and left pulmonary artery are illustrated in Figure 1.

However, as the catheter is passed toward the smaller pulmonary radicals, oxygenated blood appears either from collateral bronchial channels or from the pulmonary capillaries themselves. Such pure "pulmonary capillary blood" usually has oxygen content about 1 volume per cent higher than does peripheral arterial blood.[4]

II. ABNORMALITIES—ACYANOTIC CONGENITAL HEART DISEASE

1. *Transposed Pulmonary Veins*

Occasionally pulmonary veins empty directly into the superior vena cava and/or right atrium. Such an abnormality must be carefully distinguished from an atrial septal defect. Completely oxygenated blood under low, nonpulsatile pressure is present. It must be shown that the catheter has passed from superior vena cava into transposed pulmonary vein or from right atrium into transposed pulmonary vein without having passed through the left atrium.

2. *Atrial Septal Defect*

In atrial septal defect, a complete diagnosis can be made only by demonstrating passage of the catheter through the defect. Ideally, a double lumen catheter should be used and simultaneous pressure and oxygen value determinations obtained from right and left atrium. The tip of the catheter in the left atrium should record a slightly higher pressure than that in the right atrium; and from the tip in the left atrium, fully oxygenated blood is withdrawn. The proximal barrel of the double lumen catheter records the lower right atrial pressure curve and a lower "venous" oxygen content. Ordinarily, in atrial septal defect, right atrial oxygen content should be more than 2 volumes per cent above that of superior vena cava blood. It is important that the atrium be "explored" by means of multiple sampling of oxygen content.

Such data apply to isolated atrial septal defects in which the predominant shunt is from left to right. In the presence of pulmonary stenosis or other causes of right ventricular hypertrophy, where the principal intracardiac shunt is from right to left, and the tricuspid valve is incompetent, oxygen gradients may not be as striking. Superior vena cava oxygen may not differ appreciably from right atrial oxygen, and left atrial oxygen content may be lower than normal.[5]

PRESUMPTIVE INTERATRIAL SEPTAL DEFECT

CASE 1: A 27-year-old white female was referred for cardiac catheterization to confirm a diagnosis of interatrial septal defect. The growth and development of the patient had been normal. In 1943, while in the Navy, she had an attack of rheumatic fever and a heart murmur was first detected at that time.

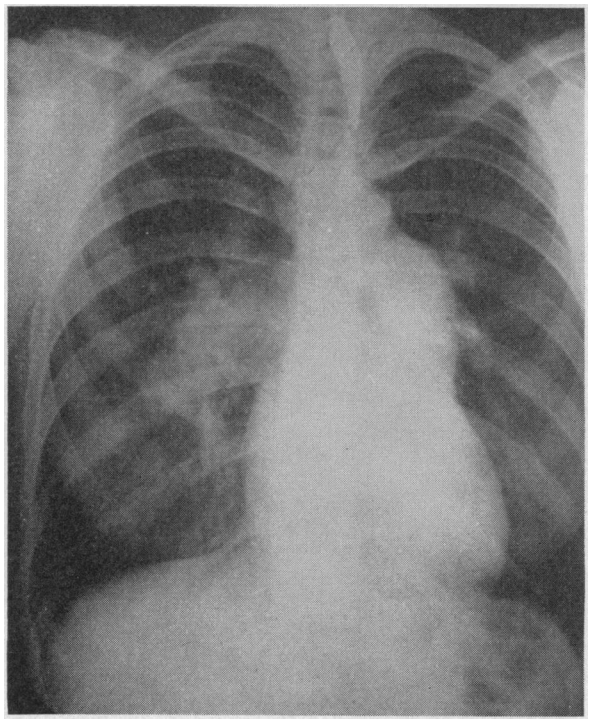

Figure 2, *A*

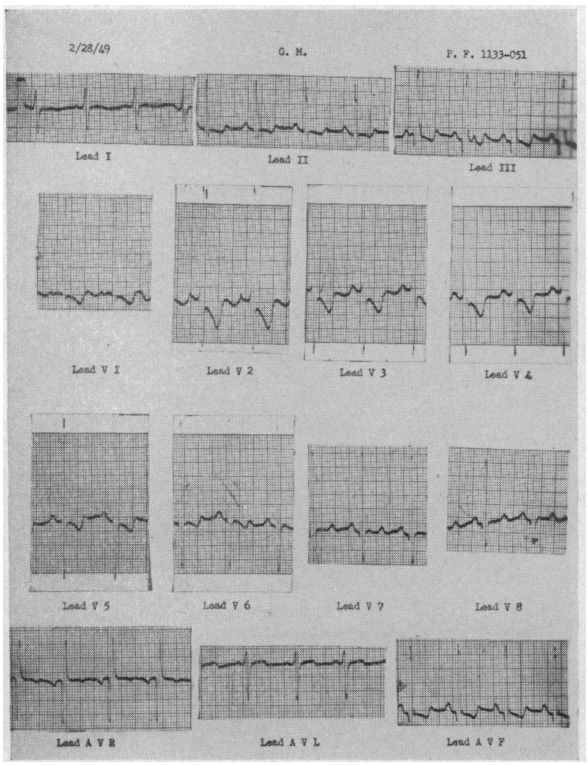

Figure 2, *B*

The patient was well developed, appeared to be well nourished and was in no acute distress. The temperature was 98.5° F., the pulse rate was 78 per minute, and the respiratory rate 20 per minute. The lungs were clear. Grade I cardiac enlargement was present. The point of maximum impulse was in the fifth intercostal space, 1 cm. outside the mid-clavicular line. P_2 and M_1 were accentuated. There was a grade V high-pitched blowing diastolic murmur, and a grade II systolic murmur at the pulmonary area.

In an orthocardiogram (Figure 2, *A*) the pulmonary arc was prominent. Pronounced dilation of the right pulmonary artery was noted in the right oblique position. The pulmonary arteries were pulsating. Right ventricular enlargement was present.

An electrocardiogram (Figure 2, *B*) gave evidence of right ventricular hypertrophy.

The hemoglobin content was 15.3 gm. per 100 cc.

Results of the cardiac catheterization are recorded in Table 1.

The diagnosis of interatrial septal defect in this patient was presumptive because of (1) an increase

TABLE 1.—*Patent Interatrial Septal Defect*

Station	Mean Pressure (mm. of Hg.)	Oxygen (Vols. %)	Oxygen Saturation (Per Cent)
1. Bifurcation of the pulmonary artery	59	14.45	69.6
2. Right ventricle	34	14.20	68.5
3. Right atrium	1 – 2	15.40	74.2
4. Superior vena cava	1 – 2	11.60	55.8
5. Left atrium	3 – 4	18.25	87.9
6. Left pulmonary vein	None	18.55	89.3

Note: With hemoglobin 15.3 gm. per 100 cc., 20.7 vol. % O_2 the saturation would be 100%.

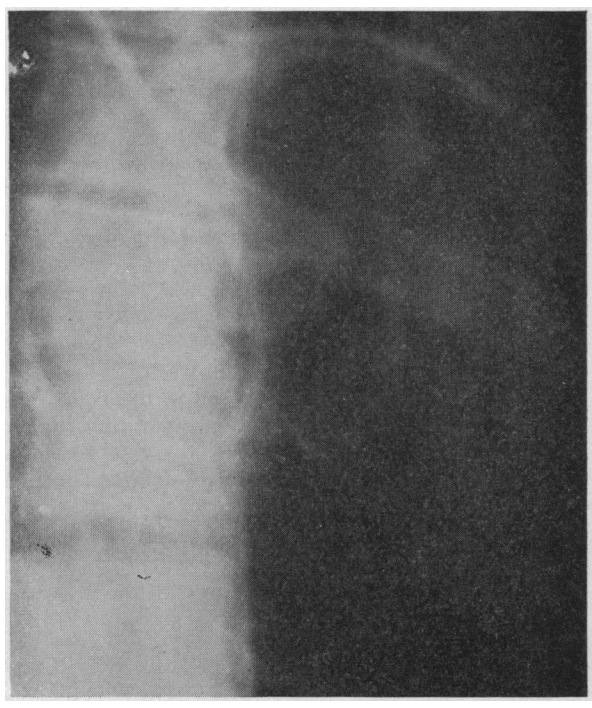

Figure 2, *C*

of oxygen content from 11.5 volumes per cent in the superior vena cava to 15.4 volumes per cent in the right auricle, and (2) because it was presumed that the catheter passed through the defect into the left auricle and out into a pulmonary vein (Figure 2, *C*). To rule out transposed pulmonary vein would have necessitated simultaneous pressure and oxygen recordings with the double lumen catheter in two positions, right atrium-left atrium, and left atrium-pulmonary vein.

3. *Ventricular Septal Defect*

An increase in oxygen content of greater than 1 volume per cent between right atrium and right ventricle is indicative of ventricular septal defect. The ventricular pressure and oxygen content depend upon (1) the size of the defect present, and (2) the proximity of the catheter to the defect itself. Occasionally the tip of the catheter may be passed through the defect, and left ventricular pressure curves may be recorded together with the sampling of fully oxygenated blood.

HIGH INTERVENTRICULAR SEPTAL DEFECT

CASE 2: A 21-year-old Japanese-American female with a history of heart disease since birth was admitted for cardiac catheterization. The only complaint was of dyspnea on exertion.

The patient was underweight. There was no dyspnea, orthopnea, cyanosis or clubbing. The pulse rate was 72 per minute. The blood pressure was 110 mm. of mercury systolic and 70 mm. diastolic. The lungs were clear. In examination of the heart the point of maximum intensity was noted in the fifth interspace at the anterior axillary line. There was a prominent systolic thrill at the second and third intercostal spaces to the left of the sternum. A grade IV systolic murmur was heard at the pulmonic area and it was transmitted to the left clavicle. A grade II pulmonary diastolic murmur was also present. P_2 was accentuated.

An orthocardiogram (Figure 3, *A*) showed that the vascular markings in both lung fields were increased, and that pronounced pulsations of the pulmonary arteries were present. The pulmonary arc was prominent. In the left oblique, both right and left ventricles were enlarged and there was a small right atrial shelf. In the right oblique the left atrium was enlarged.

An electrocardiogram showed right axis deviation (Figure 3, *B*).

Prior to catheterization, this patient was thought to have either an interatrial septal defect or a high interventricular septal defect. The results of the cardiac catheterization are recorded in Table 2.

The oxygen content of the blood in the right ventricle was approximately 2 volumes per cent higher than that of the right atrium. This established a diagnosis of interventricular septal defect. The fact

TABLE 2.—*Interventricular Septal Defect*

Station	Oxygen Content (Vols. %)	Mean Pressure (mm. of Mercury)
1. Coronary sinus	4.30	9
2. Pulmonary conus	15.70	30
3. Right ventricle	15.80	41
4. Right atrium	13.70	0
5. Superior vena cava	12.90	0
6. Femoral artery	17.45	—

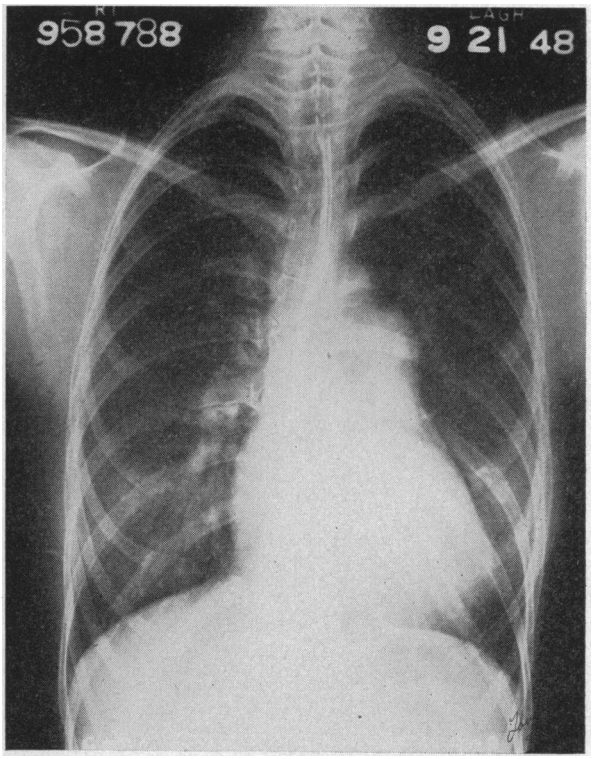

Figure 3, *A*

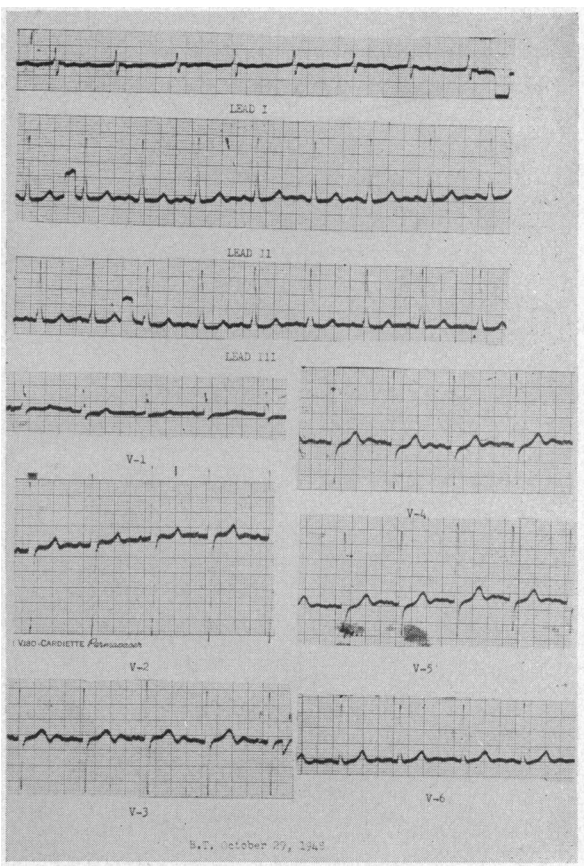

Figure 3, *B*

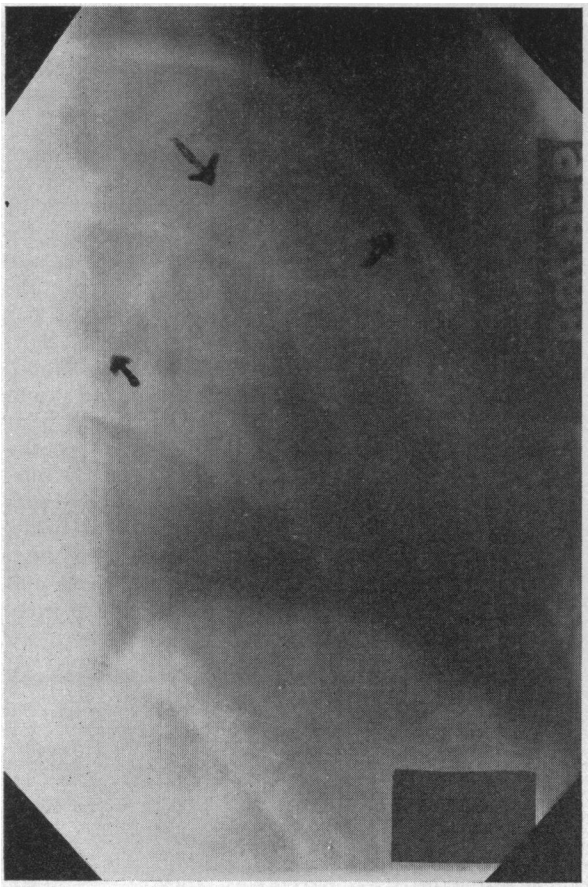

Figure 3, *C*

that the oxygen content of the blood in the right atrium was 0.8 volume per cent higher than that of the superior vena cava could possibly represent a very small interatrial septal defect. The catheter was fortuitously passed into the coronary sinus (Figure 3, *C*), and there the oxygen content was 4.3 volumes per cent. The increase in pressure in the right ventricle was secondary to the pulmonary hypertension.

4. *Patent Ductus Arteriosus*

An increase in oxygen content greater than 0.5 volume per cent between right ventricle and main pulmonary artery is indicative of some type of aortic-pulmonary shunt, usually patent ductus arteriosus. It also may be indicative of a shunt between left ventricle and pulmonary artery (high intraventricular septal defect, either alone or in combination with other defects). The actual form of the pressure curve and the degree of increase of oxygen content depends, as in ventricular septal defect, upon the size of the defect and the proximity of the catheter tip.

CASE 3: A 32-year-old white female with a history of heart disease was referred for cardiac catheterization to confirm the diagnosis of patent ductus arteriosus. The growth and development of the patient had been normal.

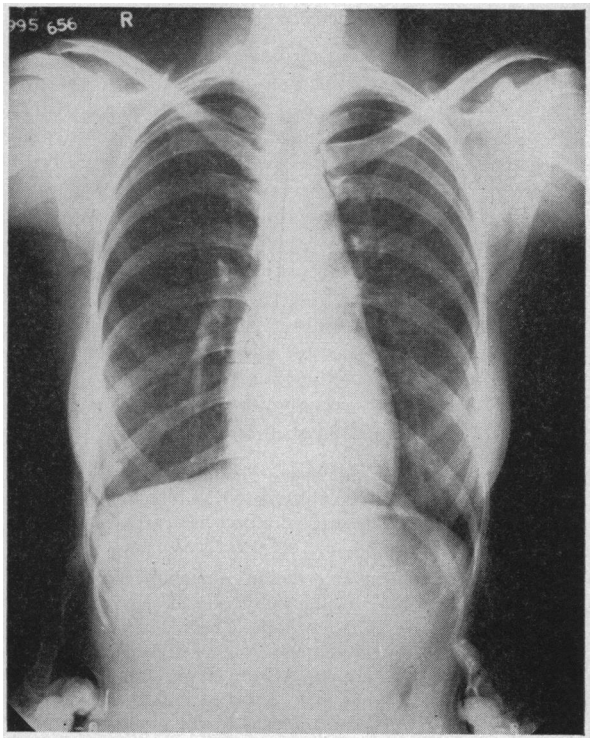

Figure 4

TABLE 3.—*Patent Ductus Arteriosus*

Station	Oxygen Content (Vols. %)	Mean Pressure (mm. of Mercury)
1. Right pulmonary artery	16	25
2. Bifurcation of main pulmonary artery	16.05	29
3. Right ventricle	14.04	16
4. Right atrium	14.5	7

The patient was thin and underdeveloped. There was no dyspnea, cyanosis or clubbing. The pulse rate was 72 per minute. The blood pressure was 115 mm. of mercury systolic and 60 mm. diastolic. The lungs were clear. A continuous "machinery" murmur over the pulmonic area was noted in examination of the heart.

An orthocardiogram (Figure 4) showed increased vascular markings, and the pulmonary arteries were described as pulsating. The heart was of normal size, and the pulmonary conus was prominent.

An electrocardiogram was normal. The hemoglobin content was 17.6 gm. per 100 cc. of blood. Results of cardiac catheterization are recorded in Table 3.

In this case, the oxygen content of the blood taken from the pulmonary artery was significantly higher (2 volumes per cent) than that in the right ventricle. This confirmed the diagnosis of patent ductus arteriosus. It is to be noted again that position of the tip of the catheter determines the oxygen and pressure values, and that the optimum position is at or near the bifurcation of the pulmonary artery. Calculation of the size of any left-to-right shunt by the usual methods may vary considerably depending upon the location of the catheter tip.[2]

5. *Isolated Pulmonary Stenosis (Non-Cyanotic)*

The diagnosis of pulmonary stenosis rests upon the finding of an abrupt rise of pulmonary systolic pressure during withdrawal of the catheter through the pulmonary valve. Ordinarily in this abnormality the pulmonary artery systolic pressure will be below 25 mm. of mercury, and the right ventricular systolic pressure considerably above that level. The diagnosis is reasonably accurate, since, under normal conditions, the systolic pressure in both pulmonary artery and right ventricle is identical. Oxygen saturation in isolated pulmonary stenosis is usually within the normal range in the peripheral arterial circulation, as the blood passing through the lungs is completely oxygenated, and there are no right-to-left intracardiac shunts.

CASE 4: A 25-year-old Negro female with a history of dyspnea on exertion since childhood and ankle edema for the previous 12 months was admitted for cardiac catheterization.

The patient appeared to be in no acute distress. The temperature was 98.6° F., the pulse rate 82 per minute, and the respiratory rate 18 per minute. The blood pressure was 132 mm. of mercury systolic and 94 mm. diastolic. The neck veins were pulsating. The lungs were clear. Grade II cardiac enlargement was present. The point of maximum intensity was in the fifth intercostal space, 3 cm. outside the midclavicular line. P_2 was accentuated. There was a grade III systolic murmur at the pulmonic area transmitted toward the left clavicle. The liver, which was palpated three fingers' breadth beneath the right costal margin, pulsated.

An orthocardiogram (Figure 5, *A*) showed marked cardiac enlargement, chiefly right ventricular, with a prominent pulmonary arc. The pulmonary vascular markings were considerably diminished.

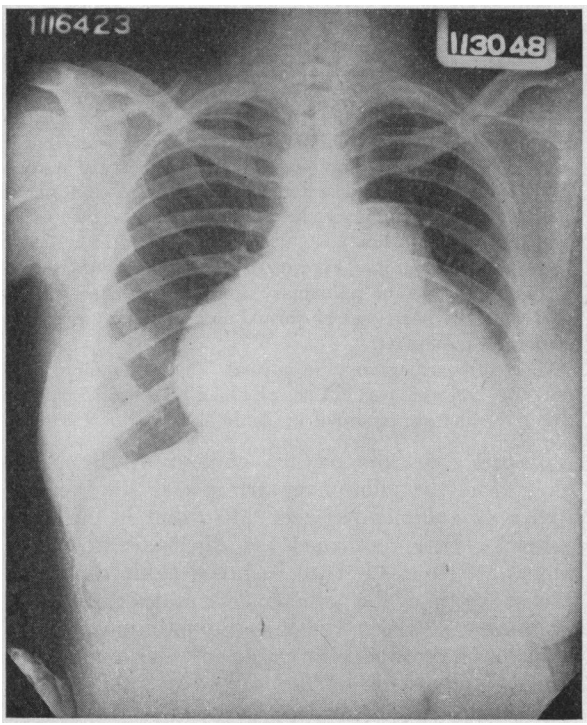

Figure 5, *A*

An electrocardiogram gave evidence of right ventricular hypertrophy (Figure 5, *B*).

Erythrocytes numbered 5.5 million, and the hemoglobin content was 17 gm. per 100 cc. The results of cardiac catheterization are recorded in Table 4.

The catheter could not be passed into the pulmonary artery. The finding of a pressure of 97 mm. of mercury in the right ventricle was consistent with the clinical and roentgenologic impression of pure pulmonary stenosis. The inability to introduce the catheter through the pulmonary valve was an additional suggestive finding. The presence of 91 per cent oxygen saturation in the femoral artery indicated complete oxygenation of pulmonary blood, and the absence of a right-to-left intracardiac shunt. These three findings—(1) starved lungs, with high right ventricular pressure, (2) the inability of the catheter to enter the pulmonary artery and (3) normal arterial oxygen saturation—established the presumptive diagnosis of pure pulmonary stenosis. Illustrative cases of pulmonary stenosis with *diagnostic* pressure gradients from right ventricle to pulmonary artery will be reported later in this presentation (Cases 5 and 6).

TABLE 4.—*Pure Pulmonary Stenosis*

Station	Mean Pressure (mm. of Hg.)	Oxygen (Vols. %)	Oxygen Saturation (Per Cent)
1. Right ventricle	97	10.37	49.1
2. Right atrium	19	9.99	48.8
3. Subclavian vein	19	9.97	46.7
4. Femoral artery		18.8	91.7

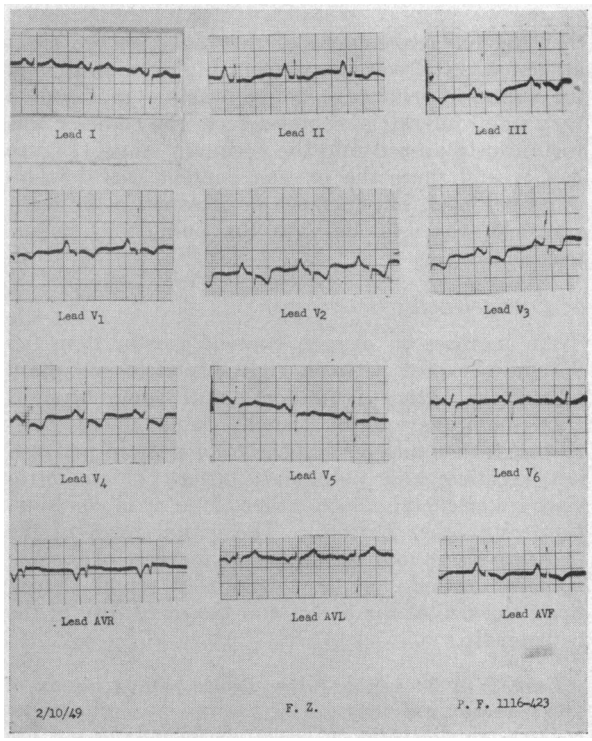

Figure 5, *B*

III. CYANOTIC CONGENITAL HEART DISEASE

1. *Tetralogy of Fallot*

The tetralogy of Fallot is the commonest type of cyanotic congenital heart disease and consists of (1) an interventricular septal defect, (2) dextroposition of the aorta, (3) right ventricular hypertrophy and (4) pulmonary stenosis.

It is not always possible, with the technique of cardiac catheterization, to arrive at a "complete" physiologic diagnosis. Nevertheless, significant and valuable information is almost invariably obtained. The data may be subdivided thus:

1. *Pulmonary stenosis.* This is the crucial determination, and, as was noted earlier, is determined best by the rise of systolic pressure during continuous withdrawal of the catheter from pulmonary artery to right ventricle. In addition, information may be gathered as to the actual presence of one or both pulmonary arteries, provided that the catheter is manipulated into the terminal portion of each pulmonary artery.

2. *Hypertrophy of the right ventricle.* This is a result of pulmonary stenosis and is evidenced by (a) elevated right ventricular systolic pressure, (b) right axis deviation with evidence of hypertrophy in the electrocardiogram, and (c) fluoroscopic demonstration of right ventricular enlargement.

3. *Dextroposition, or overriding aorta.* Four methods are available to demonstrate overriding: (a) arm-to-tongue circulation time (approximates arm-to-lung time), (b) angiocardiography will reveal the early presence of dye in the aorta when it can be definitely proven that dye has passed directly from right atrium to right ventricle to aorta, (c) the catheter itself may be seen to pass from apparent right ventricle to aorta, (d) a right ventricular systolic pressure equal to aortic systolic pressure is fairly good evidence for the presence of overriding. It is apparent that no one of the above methods is infallible, and all should be utilized during cardiac catheterization.

4. *Interventricular septal defect.* This defect in cyanotic heart disease is not simple to demonstrate by the technique of catheterization, since the shunt may be mainly from right to left. This septal defect may be assumed to be present when overriding of the aorta is demonstrated.

The differentiation of tetralogy of Fallot from the tetralogy of Eisenmenger requires, in addition, the determination of oxygen consumed per liter of ventilation and the determination of changes in arterial oxygen saturation with exercise. In the tetralogy of Fallot the arterial oxygen saturation falls with exercise, as does oxygen consumed per liter of ventilation.[1] In the tetralogy of Eisenmenger the arterial oxygen saturation falls with exercise, and the oxygen consumption per liter of ventilation rises.

CASE 5: A 21-year-old white male with a history of cyanosis and clubbing since four years of age was admitted for cardiac catheterization. The only complaint was that of dyspnea on moderate exertion.

The patient was moderately underdeveloped. Pronounced cyanosis of the skin and mucous membranes was noted. There was clubbing of the fingers, toes, and nose. The temperature was 98° F., the pulse rate 120 per minute and the respiratory rate 20 per minute. The blood pressure in the right arm was 118 mm. of mercury systolic and 90 mm. diastolic; in the left arm, 115 mm. systolic and 100 diastolic. The lungs were clear. There was a harsh grade III systolic murmur best heard over the third and fourth left interspaces and transmitted well in all directions. P_2 was louder than A_2.

An orthocardiogram showed some decrease in the peripheral lung markings. The cardiac silhouette was not enlarged. In the right oblique the pulmonary artery appeared quite small (Figure 6, *A*).

An electrocardiogram gave evidence of right axis deviation with pronounced clockwise rotation of the heart (Figure 6, *B*).

The hemoglobin content of the blood was 23 gm. per 100 cc. Erythrocytes numbered 8.5 million, and leukocytes 7,600.

The results of cardiac catheterization are recorded in Table 5. In Figure 6, *C* there are simultaneous pressure

TABLE 5.—*Tetralogy of Fallot*

Station	Oxygen Content (Vols. %)	Oxygen Saturation (Per Cent)	Pressure (mm. of Hg.) Systolic	Diastolic
1. Main pulmonary artery	17.54	48.5	25 to 33	18 to 21
2. Right ventricle (*High*)	18.7	51.7	155 to 167	28 to 32
2. Right ventricle (*Low*)	17.80	49.3		
3. Right atrium	17.65	48.8	15 to 20	5 to 10
4. Superior vena cava	19.88	55	15 to 20	5 to 10
5. Left brachial artery	25.85	71.6	90	75

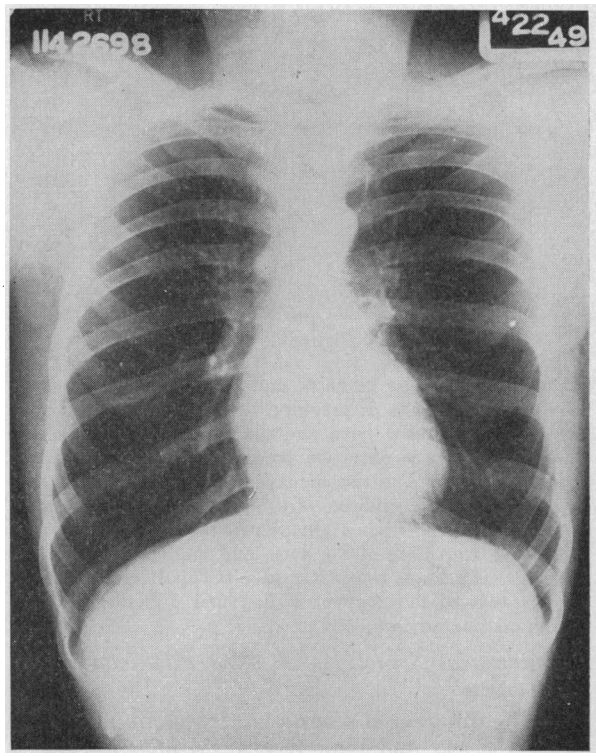

Figure 6, *A*

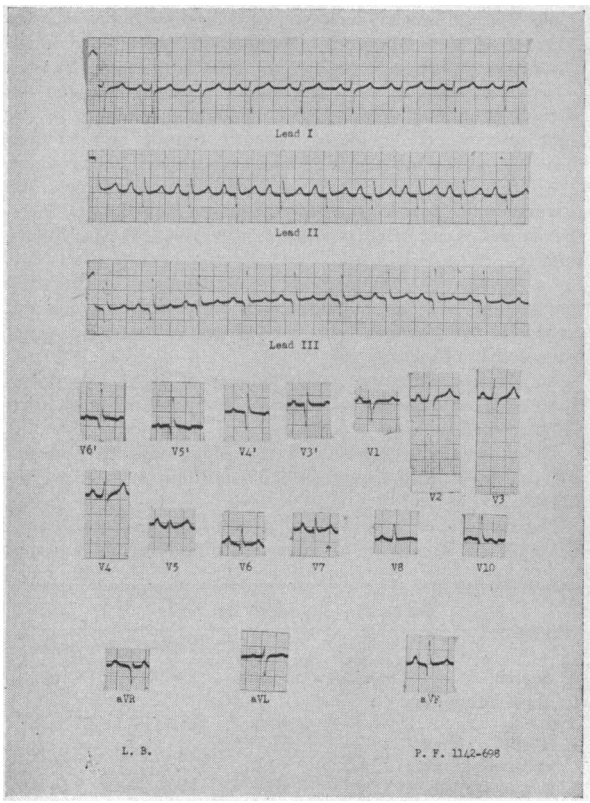

Figure 6, *B*

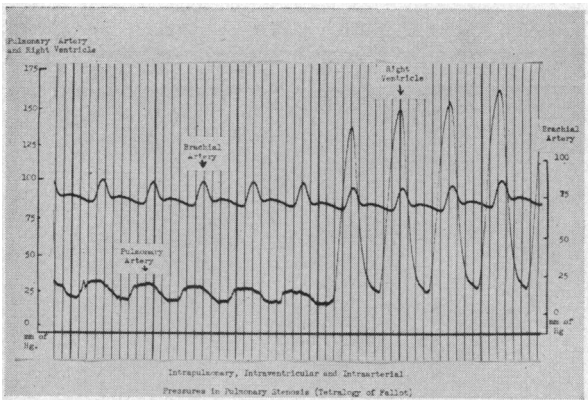

Figure 6, *C*

recordings from the catheter and an intra-arterial needle. The pronounced rise in pressure (Figure 6, *C*) as the catheter was withdrawn from the pulmonary artery into the right ventricle was objective proof of the presence of pulmonary stenosis. The rise of oxygen high in the right ventricle was possible evidence of a high interventricular septal defect. The very high right ventricular systolic pressure suggested overriding of the aorta and implied that the predominant shunt was from right to left. The diagnosis arrived at was that of tetralogy of Fallot, and a Blalock-Taussig operation was advised.

2. *Interatrial Septal Defect Plus Pulmonary Stenosis*

In the differential diagnosis of cyanotic congenital heart disease, pulmonary stenosis plus interatrial septal defect holds an important place. Operative results in this condition have been equivocal in spite of the presence of pulmonary stenosis and pronounced subnormality of oxygen content in arterial blood. To demonstrate the interatrial septal defect, the oxygen content of superior and inferior caval blood is a matter of great importance. The dynamics of the interatrial defect include shunts from right to left and from left to right. Ideally, the catheter should be passed through the interatrial defect into the left atrium.

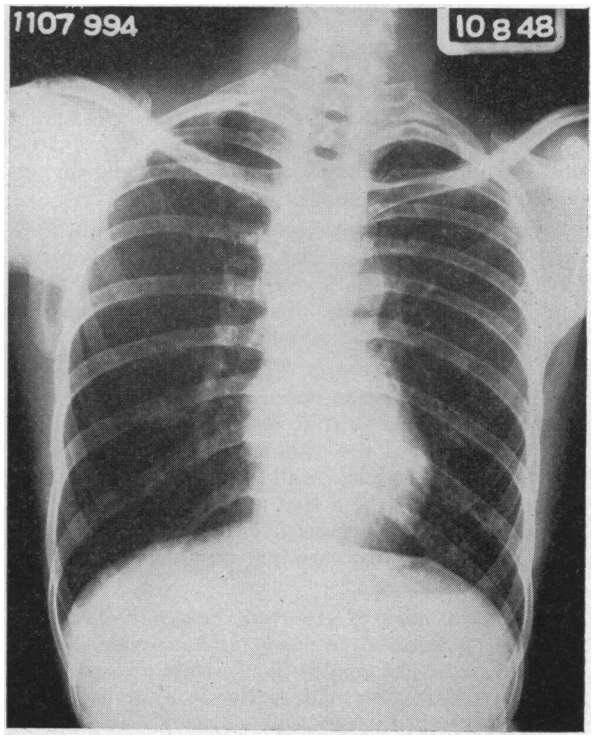

Figure 7, *A*

CASE 6: A 23-year-old white male, cyanotic since birth, who complained of dyspnea and increase of cyanosis on exertion, was admitted for cardiac catheterization. He had frequently assumed a "squatting" position in the past. Growth and development had been somewhat impaired.

Upon physical examination, clubbing of fingers and toes was noted. The pulse rate was 66 per minute. The blood pressure was 95 mm. of mercury systolic and 70 mm. diastolic. The lungs were clear. The heart was not enlarged. There was a systolic thrill at the second and third interspaces to the left of the sternum. A grade IV systolic murmur was present at the second, third and fourth intercostal spaces to the left of the sternum and was transmitted into the neck.

An orthocardiogram showed some decrease in the vascular markings in the periphery of the lung fields (Figure 7, *A*). The heart was small and there was no concavity in the region of the pulmonary arc. In the left oblique, the right ventricle appeared to make up at least two-thirds of the cardiac silhouette.

An electrocardiogram gave evidence of right ventricular hypertrophy (Figure 7, *B*).

The hemoglobin content of the blood was 27.5 gm. per 100 cc. The results of cardiac catheterization are recorded in Table 6.

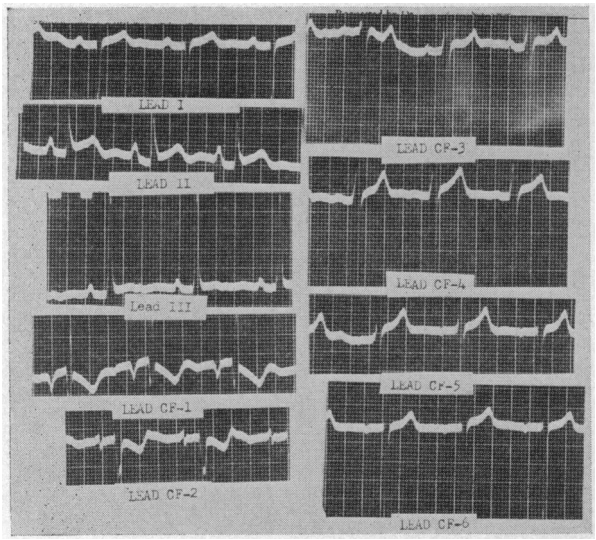

Figure 7, *B*

TABLE 6.—*Pulmonary Stenosis with Either an Interatrial Septal Defect or Transposed Pulmonary Veins*

Station	Oxygen Content (Vols. %)	Oxygen Saturation (Per Cent)	Mean Pressure (mm. of Mercury)
1. Right pulmonary artery	21.7	58.5	2 – 3
2. Right ventricle	22.1	59.6	39 – 61
3. Right atrium	22.1	59.6	1 – 2
4. Superior vena cava	18.5	50.0	0
5. Femoral artery	25	63.8	—

Note: With hemoglobin content at 27.5 gm. per 100 cc., 37.04 volumes % of oxygen would be needed for 100% saturation.

The oxygen content of the right atrium was 3.6 volumes per cent higher than that of the superior vena cava, representing either interatrial septal defect or transposed pulmonary veins. Also, the pressure, which was elevated in the right ventricle (39 to 61 mm. of mercury), dropped to 3 to 4 mm. in the pulmonary artery. From these data, the diagnosis of pulmonary stenosis was proven.

3. *Tetralogy of Eisenmenger*

The main diagnostic differential from tetralogy of Fallot is the tetralogy of Eisenmenger in which is found (1) a high interventricular septal defect, (2) right ventricular hypertrophy, (3) dextroposition of the aorta and (4) a *normal* or *dilated* pulmonary artery. Cyanosis usually does not appear until about the age of puberty.

In the complex of Eisenmenger, the following determinations should pertain: (1) identical and simultaneous systolic pressure in pulmonary artery and right ventricle (both elevated above the normal, usually by a considerable amount). Both should be identical, or very nearly identical with that of aortic systolic pressure. (2) Right ventricular hypertrophy is self-evident in the above. (3) The interventricular septal defect is more often evident than in the tetralogy of Fallot, and thus, right ventricular oxygen content tends to be higher than right

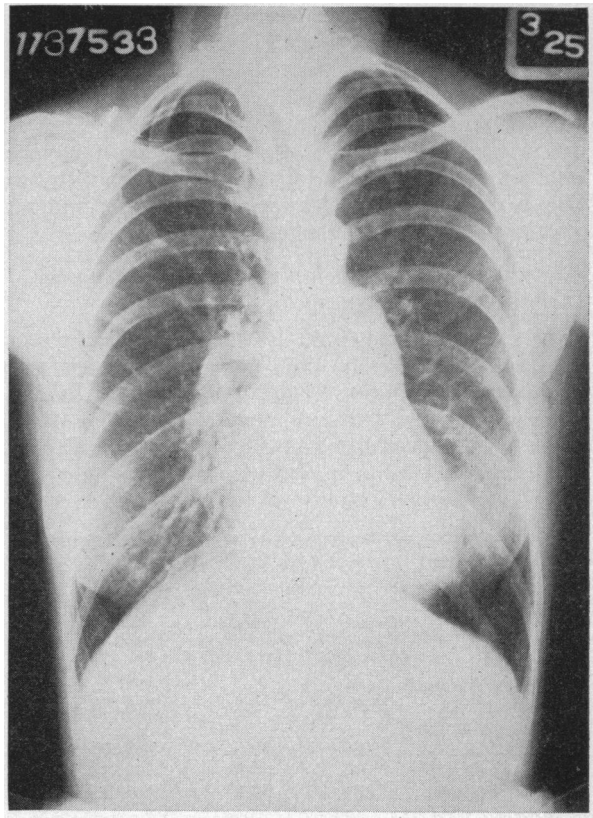

Figure 8, *A*

atrial oxygen content. (4) Finally, although there is a fall of arterial oxygen saturation with exercise as in the tetralogy of Fallot, the oxygen consumed per liter of ventilation rises.

CASE 7: A 29-year-old white female, cyanotic since age 13, was admitted for cardiac catheterization. The cyanosis had persisted since onset, and clubbing of the fingers and toes had developed. Dyspnea occurred only on severe exertion.

Upon physical examination, cyanosis of the skin and mucous membranes was noted. Clubbing of fingers and toes was present. The temperature was 99° F., the pulse rate was 84 per minute, and the respiratory rate was 18 per minute. The blood pressure was 100 mm. of mercury systolic and 78 mm. diastolic. The lungs were clear. The heart was not enlarged on percussion. The point of maximum intensity was in the fifth intercostal space at the midclavicular line. P_2 was louder than A_2. There was a grade II systolic murmur at the base, best heard over the pulmonic area, and transmitted into the neck and toward the apex.

An orthocardiogram showed increased pulmonary markings with pulsating pulmonary arteries. Grade II right ventricular enlargement was present (Figure 8, *A*). An electrocardiogram gave evidence of right ventricular hypertrophy (Figure 8, *B*). Hemoglobin content of the blood was 19.5 gm. per 100 cc. Leukocytes numbered 7,700. The arm-to-tongue circulation time (dehydrocholic acid) was 12 seconds, and the arm-to-lung circulation time (ether) was 8 seconds.

The results of cardiac catheterization are recorded in Table 7, and the pressure readings are recorded in Figure 8, *C*.

The pressure in the main pulmonary artery was 102 mm. of mercury systolic and 50 mm. diastolic; pressure in the right ventricle was 99 mm. systolic

and 2 mm. diastolic. The following measurements were pertinent and typical: (1) The right ventricular systolic pressure, and the pulmonary artery systolic pressure were considerably elevated and similar. (2) The pressures in both approximated the brachial artery systolic pressure. (3) The high oxygen content of pulmonary artery was presumably caused by a high interventricular septal defect. The findings were typical of Eisenmenger's complex.

4. *Rudimentary Right Ventricle and Transposed Pulmonary Veins*

In cyanotic congenital heart disease, right axis deviation is definitely more common than left axis deviation. The finding of left axis deviation usually suggests a small or rudimentary right ventricle, often with the added feature of tricuspid atresia. (Left axis deviation may also occur in single ventricle or with a tetralogy of Fallot with dextrocardia.) The following case illustrates these features and, in addition, the presence of pulmonary veins entering the right atrium.

TABLE 7.—*Tetralogy of Eisenmenger*

Station	Oxygen Content (Vols. %)	Oxygen Saturation (Per Cent)	Pressure Syst./Diast. (mm. Hg.)	Pressure Mean
1. Right pulmonary artery	17.8	76.5	115/50	78–82
2. Main pulmonary artery	17.6	70.6	102/50	78–80
3. Right ventricle	16.0	60.6	99/2	43–60
4. Right atrium	15.5	58.8	6/2	2–4
5. Superior vena cava	14.7	55.7		
6. Right brachial artery	21.8	82.7	115/75	95

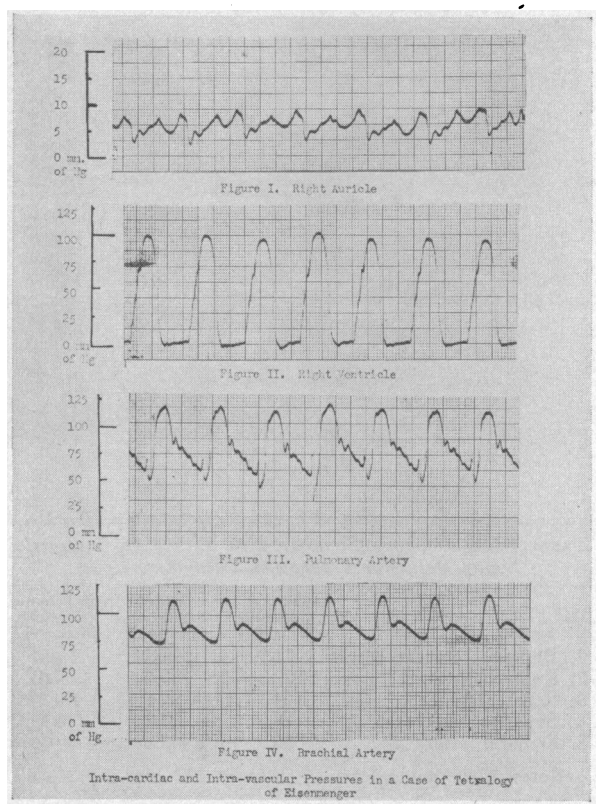

Figure 8, *C*

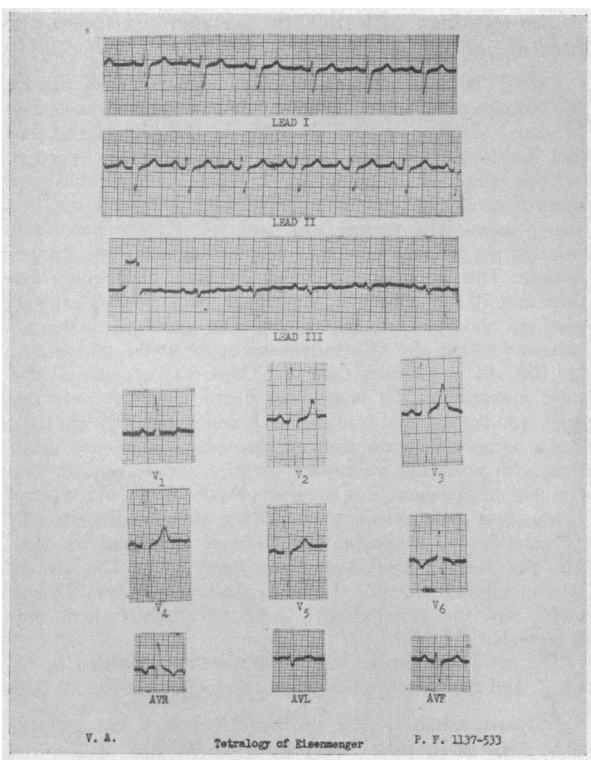

Figure 8, *B*

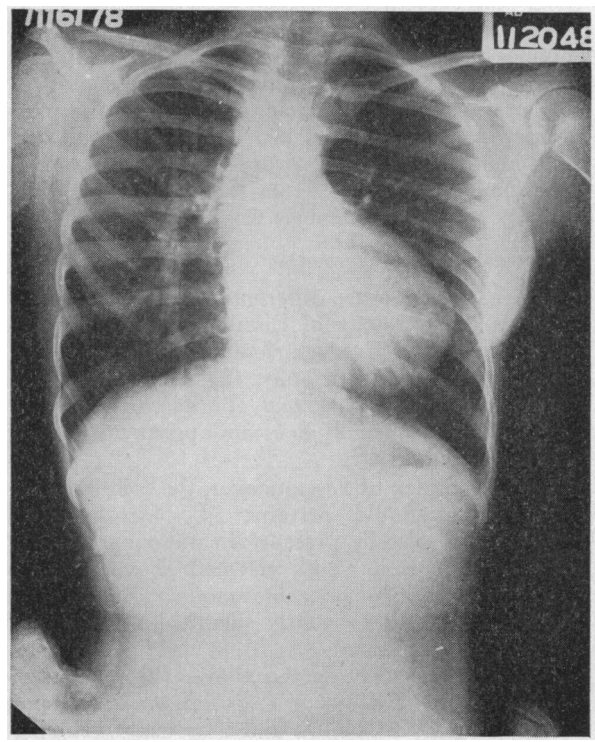

Figure 9, *A*

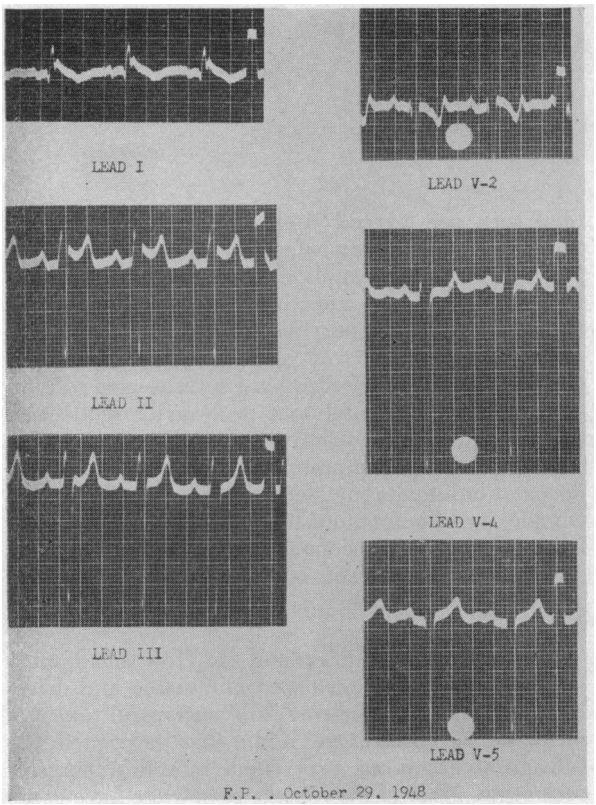

Figure 9, *B*

CASE 8: A 13-year-old white female, a "blue baby," and cyanotic since birth, was admitted to the Los Angeles County Hospital on November 20, 1948, for cardiac catheterization. Growth and development had been normal, but the exercise tolerance had been low as long as the patient could remember.

The patient was well developed. Cyanosis and clubbing of fingers and toes were present. The blood pressure was 94 mm. of mercury systolic and 88 mm. diastolic. The lungs were clear. Upon examination of the heart the point of maximum intensity was located in the fifth intercostal space at the midclavicular line. P_2 was accentuated and greater than A_2. There was a grade III systolic murmur in the fifth interspace just inside the midclavicular line.

An orthocardiogram (Figure 9, *A*) showed a boot-shaped left ventricle in the posterior-anterior view. One plus enlargement was present. In the left oblique view there was a straightening of the normal contour of the right ventricle which suggested a small or non-functioning right ventricle.

An electrocardiogram gave evidence of pronounced left axis deviation. The QRS interval was prolonged (Figure 9, *B*).

The results of cardiac catheterization are recorded in Table 8. Prior to catheterization, the main features of this case were: (1) cyanotic heart disease with left axis deviation and (2) an orthocardiogram showing a small or rudimentary right ventricle.

TABLE 8.—*Rudimentary Right Ventricle with Transposition of the Pulmonary Veins*

Station	Oxygen Content (Vols. %)	Oxygen Saturation (Per Cent)	Mean Pressure (mm. of Mercury)
1. Right pulmonary vein	24.5	80.0	9
2. Coronary sinus	4.45	14.5	40
3. Right atrium	16.8	53.4	18
4. Superior vena cava	14.7	46.8	9
5. Aorta	20.4	65.3	40

During the catheterization, the catheter was first observed to pass directly from the superior vena cava and the right atrium into the right pulmonary vein, from which oxygenated blood was obtained. Later, the catheter was passed into the aorta (presumably from right ventricle, through interventricular septal defect, into left ventricle) and appeared in the descending aorta at about the level of the second lumbar vertebra.

Reviewing all the findings, the following defects were established: (1) transposed pulmonary veins (by the presence of completely oxygenated blood in a vein emptying into the superior vena cava); (2) rudimentary or nonfunctioning right ventricle (straight anterior margin of the heart in the left oblique position by x-ray and the extreme left axis deviation in the electrocardiogram. It is presumed (but not proven) that interatrial septal defect, with or without interventricular septal defect, completes the picture in this malformation.

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REFERENCES

1. Bing, R. J., Handelsman, J. C., and Campbell, J. A.: Physiologic diagnostic tests in congenital heart disease, Modern Concepts Cardiovas. Disease, 17: [n.p.], March 1948.

2. Cournand, A., Baldwin, J. S., and Himmelstein, A.: Cardiac Catheterization in Congenital Heart Disease. Commonwealth Fund. New York City. 1949.

3. Cournand, A., and Ranges, H. A.: Catheterization of right auricle in man, Proc. Soc. Exp. Biol. and Med., 46:462, March 1941.

4. Dexter, L., Haynes, F., Burwell, C., Eppinger, E., Sagerson, R., and Evans, J.: Studies of congenital heart disease. II. The pressure and oxygen content of blood in the right auricle, right ventricle and pulmonary artery in control patients, with observations on the oxygen saturation and source of pulmonary "capillary" blood, J. Clin. Investigation, 26:554, 1947.

5. Vandam, L. V., Bing, R. J., and Gray, F. D., Jr.: Physiological studies in congenital heart disease. IV. Measurements of the circulation in five selected cases, Bull. J. Hopkins Hosp., 81:192, Sept. 1947.